Diffuse Cystic Lung Disease

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Learning objectives

1 - Review the definition of cyst on lung parenchyma.

2 - Explain the differential diagnosis of entities which present themselves with diffuse lung cysts.

3 - To describe and illustrate the HRCT finding of these entities.
Background

A cyst is a thin-walled (less than 2-3 mm in thick), well-defined, air-containing lung lesion 1 cm or more in diameter. When assessing lung cysts on HRCT, the most important feature that allows differentiation between diseases is their distribution. Careful scrutiny of cyst shape and size, as well as for the presence of any ancillary findings, may help refine the differential diagnosis.

Pulmonary cysts can be found in many entities, like emphysema, pneumonia, cystic bronchiectasis and subacute hypersensitivity pneumonitis. However few diseases are characterized by cysts as their primary abnormality.

These are rare, and include Langerhans cell histiocytosis (LCH), lymphangiomyomatosis (LAM), tuberous sclerosis (TS), neurofibromatosis, lymphoid interstitial pneumonia (LIP) and Sjoren's syndrome.

LCH and LAM are the most common to present with diffuse lung cysts.
Imaging findings OR Procedure details

Pulmonary Langerhans Cell Histiocytosis (LCH)

Most patients with pulmonary LCH are young or middle-aged adults (between the ages of 20 and 40 years), and over 90% of whom are cigarette smokers. Lung involvement in LCH is common, seen in 40% of patients, and may be an isolated abnormality.

The HRCT appearances of LCH vary according to the chronicity of the disease. In the early stages, nodules (which correspond with Langerhans cell granulomas) are the predominant features, while cysts tend to develop later.

In some patients, cysts are the only abnormality visible on HRCT, but in many cases, small nodules (usually less than 5 mm in diameter) are also present. Some nodules may show lucent centers, presumably corresponding to small "cavities".

In almost all patients, HRCT demonstrates cystic air spaces, which are usually less than 10 mm in diameter. Although many cysts appear round, they can also have bizarre shapes, being bilobed or cloved-leaf in appearance. An upper lobe predominance in the size and number of cysts is common, typically sparing the lung bases and the costophrenic angles.

LCH lesions typically are peribronchiolar in distribution.

Associated hilar or mediastinal lymph node enlargement or lytic bone lesions also may be present.

Lymphangioleiomyomatosis (LAM)

LAM occurs almost exclusively in women of childbearing age, usually between 17 and 50 years.

On HRCT, patients with LAM characteristically show numerous isolated, thin-walled, rounded lung cysts. They are most often distributed diffusely throughout the lungs, from apex to base, and no lung zone is spared (perhaps the most useful differentiating sign between LCH and LAM). The size of the cysts tend to increase with progression of the disease.

Patchy areas of ground-glass opacity sometimes are also seen and they may represent areas of pulmonary hemorrhage.

Spontaneous or recurrent pneumothorax may be the presenting finding in up to 50% of patients. Chylous pleural effusions can also be present.
LAM is now considered an indication for lung transplantation. The abnormality may recur in the transplanted lung.

**Lymphoid Interstitial Pneumonia (LIP) and Sjögren's Syndrome**

LIP often occurs in association with collagen-vascular diseases and Sjögren's syndrome. Ground-glass opacities and nodules are almost universal features in LIP, with cysts seen in about two-thirds of patients.

Cysts are most typical of LIP in Sjögren's syndrome. They tend to be fewer in number than those seen in patients with LCH or LAM.

**Cystic Pulmonary Metastatic Disease**

Cystic pulmonary metastases occur most frequently in tumors of epithelial origin. The appropriate history of primary malignancy is critical to suggest this diagnosis.

**Sub-acute hypersensitivity pneumonitis (HP) and Pneumocystis Jiroveci Pneumonia (PJP)**

Cysts on HRCT are seen in approximately 10% of patients with subacute HP and are usually few in number and random in distribution. Despite their low frequency, the presence of cysts can be a helpful clue in making a radiological diagnosis of HP, when identified in conjunction with the more classical signs of the disease: these are centrilobular ground glass nodules and a mosaic attenuation pattern.

Differentiating between HP and PCP is based on clinical history and imaging features. PCP in patients with AIDS commonly presents with diffuse ground glass opacification, sepal thickening and occasional cysts, but with no small airways disease.

**Centrilobular emphysema (CLE)**

Areas of CLE are typically not surrounded by a wall, however, CLE may appear as thin walled cysts and on occasion can be very difficult to distinguish for other entities. Looking for certain CT features of CLE, like the presence of multiple rounded areas of low attenuation or a central core vessel (centrilobular artery) in the low attenuation cystic space, can help.
Images for this section:

**Fig. 1**: Pulmonary Langerhans Cell Histiocytosis (LCH) - Uncountable bizarre shaped cysts, with relative sparing of costophrenic angles.

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**Fig. 2**: Lymphangioleiomyomatosis (LAM) - Round cysts uniformly distributed through lung parenchyma. Pneumothorax is present (arrow contour).

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Fig. 3: Lymphoid Interstitial Pneumonia (LIP) - Patient with Sjögren syndrome diagnosis with ground glass opacities (arrow contour) associated with few cysts (arrow)

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Fig. 4: Cystic Pulmonary Metastatic Disease - In a background history of squamous cell carcinoma, many metastatic lesions are seen on lung parenchyma, some of them with a cystic appearance (arrow).

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Fig. 5: Sub-acute hypersensitivity pneumonitis (HP) - A mosaic attenuation pattern associated with cysts is present. Few small centrilobular nodules can also be seen.

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Fig. 6: Pneumocystis Jiroveci Pneumonia (PJP) - Patient with AIDS present with diffuse ground glass opacification, minimal septal thickening and few cysts. Clinical history supported the diagnosis.

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**Fig. 7:** Emphysema - Diffuse low attenuation lung parenchyma with areas not supported by a wall (arrow contour). Few thin walled cysts are seen (arrow).

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Conclusion

Lung cysts on HRCT are not an infrequent finding and require explanation where possible. By scrutinizing the distribution and shape of pulmonary cysts as well as looking for ancillary CT signs, in certain conditions HRCT is diagnostic and no further investigation is required. Although LAM and LCH are the most frequently encountered causes of thin walled cysts at HRCT, there are other entities that should be considered.
References